REFRESHER COURSE FOR GENERAL PRACTITIONERS

CONGENITAL HEART DISEASE—I

BY

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The exact diagnosis of many forms of congenital heart disease is difficult and may require complicated investigations. Some forms, however, can be diagnosed with confidence by simple clinical methods, and most others can be placed within a group that allows good advice to be given about the degree of activity and the prognosis. Symptoms are generally more important than physical signs in judging this. Too many patients are still more limited than they should be because of the finding of a systolic murmur; the improvement that is sometimes noted after the age of 14 may simply mean that the child has broken free from unnecessary restrictions imposed at school, and has increased his activity within the natural limitations of his heart.

The general principle should be to find the child's full capacity by a gradual increase in activity and then to keep the daily activity a little below this, with adequate rest and a long night in bed. The child should be carefully treated for transient infections, and sulphonamides should be used readily to minimize the risks of bacterial endocarditis.

The first fundamental division of congenital heart disease is into acyanotic and cyanotic cases. This is so important that the cyanotic group should mean indubitable cyanosis at rest, and those in whom cyanosis is only occasional or seen only sometimes after exertion should be left in a special third group.

FIG. 1.—The three common causes of left-to-right shunts. If large enough these will cause pleonaemic lungs.

Central cyanosis at rest indicates a right-to-left (venous-arterial) shunt and therefore gives at once a partial diagnosis. Senac as long ago as 1749, in his *Traité de la structure du cœur, de son action et de ses maladies*, explained cyanosis as being due to a mixture of venous with arterial blood through an absence of the ventricular septum, but it is only recently that the full validity of this generalization has been accepted (Fig. 1).

The second division, which has become almost as fundamental since the possibility of increasing the blood flow to the lungs by subclavian-pulmonary anastomosis (Blalock-Taussig operation), is into those in whom the blood flow to the lungs is increased and those in whom it is diminished. Although this is of more practical importance for cyanotic cases, it provides a helpful subdivision for acyanotic cases also. The distinction is made mainly by radioscopy, but in well-marked cases the radiograph may be distinctive and on physical examination the increase or diminution of the pulmonary second sound may give a clue to which is likely to be found (Figs. 2a and 2b).

The distinction is so important that I have introduced the words "oligaemic" and "pleonaemic" to indicate the groups where the blood flow to the lungs is diminished and increased respectively* (Fig. 3). Oligaemic lungs are shown by light lung fields, often with light vascular markings. They are most common in the cyanotic group, though acyanotic

FIG. 2A.—Pleonaemic lungs.

FIG. 2B.—Oligoemic lungs.

examples are found in pure pulmonary valvular stenosis. Pleonaemic lungs are shown by dense lung fields, generally with large pulmonary arteries and increased vascular markings, often with easily seen pulsation in the pulmonary branches.

*I am indebted to Mr. E. L. York, classical tutor of New College, Oxford, for *ολιγαίμος*, and to Dr. H. St. H. Vertue for the companion word *πλεοναίμος*.

Pleonaemic lungs indicate a left-to-right (arteriovenous) shunt and so give part of the diagnosis at once. Among the acyanotic cases auricular septal defect, ventricular septal defect, and patent ductus arteriosus are the main examples, and all these may give a somewhat similar radio-

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FIG. 3.

graph, though in most cases of ventricular septal defect the opening is small ("maladie de Roger") and the changes in the lung fields may then be trivial.

The same radiological picture in cyanotic cases indicates that there is a left-to-right shunt, as well as the right-to-left shunt causing the cyanosis.

ACYANOTIC CONGENITAL HEART DISEASE

In the acyanotic group two conditions are of special importance because they can be cured by surgery, and both can be diagnosed by simple clinical methods—patent ductus arteriosus and coarctation of the aorta. In the others exact diagnosis is less important, and advice should be based mainly on the child's capacity, though naturally the size of the heart will influence this.

Ventricular and Auricular Septal Defect

Ventricular septal defect is thought to be the commonest congenital lesion. Occasionally the defect may be large and high, and then the mortality is considerable even in infancy; more often it is small ("maladie de Roger"), and this is perhaps the least serious of all congenital lesions, many patients leading a normal life as adults. A rough systolic murmur in the fourth space on the left side with a thrill in a patient with little disability may be taken as good evidence of this diagnosis provided there are no other gross abnormal findings.

Auricular septal defect is most often recognized for the first time in adult life (70% between 20 and 50 years of age). It is the most common congenital lesion found *post mortem*, though the "maladie de Roger" is probably more common. The most distinctive feature is the x-ray appearance of the pleonaemic lung in its most complete form, with large dilated pulmonary arteries with striking pulsation, though a true "hilar dance" is seen in less than half.

Clinically, the main characteristic is the large heart with a good capacity for exertion, though, as most patients finally die with congestive failure, this to some extent depends on the stage at which the diagnosis is made. Two-thirds of the patients have a systolic murmur in the pulmonary area and often at the apex also, and nearly a quarter have a pulmonary diastolic murmur. The pulmonary second sound is accentuated in more than half of these cases.

The electrocardiogram is helpful: right axis deviation is present in 80%, some widening of QRS above 0.1 second in 50%, and a P-R interval of over 0.2 second in about 40%, often with a large bifid P wave.

Calculations from the relative size of the aorta and pulmonary artery in average cases of auricular septal defect suggest that half the volume of blood in the left auricle is shunted into the right auricle, and that two-thirds of the blood is circulating round the pulmonary circuit and one-third round the systemic circuit.

Towards the end, as failure is developing, many patients get slight cyanosis on exertion, but not much more than patients with mitral stenosis. Haemoptysis may occur; and heart failure and, less commonly, pulmonary infarction are the main causes of death.

Patent Ductus Arteriosus

This lesion can be diagnosed by a continuous murmur though systole and diastole, generally best heard on the left side in the first and second spaces and sometimes very high up under the clavicle. The thrill may be equally continuous or may be mainly in systole. With this there is generally a wide pulse pressure and other signs such as are found in aortic incompetence, and the degree of these will give some idea of the amount of blood passing through the ductus. The size of the heart and the degree of dilatation of the pulmonary artery on radioscopy will provide further evidence. Often the continuous murmur may not develop till the child is 3 or 4 years old, and before this, in the presence of a systolic murmur only, no certain conclusion can be reached without full investigations, which are not indicated at this stage.

This condition can be cured completely by operation, and, if symptoms or significant enlargement of the heart make operation advisable, it should be carried out between the ages of 6 and 12 or, indeed, at any time up to the age of 20. Even after 20 the advantages of operation should not be forgone in suitable cases, but the risk is greater and the disability may not justify it. Some authorities advise operation in every case in which the diagnosis is made with certainty, but in early childhood there is some chance of the ductus closing spontaneously, and many patients with little enlargement of the heart get on well through adult life. Bacterial endocarditis is a considerable risk, but now that it can be cured with penicillin this danger is a less decisive indication for immediate operation.

The risk of operation is not great in the hands of an experienced thoracic surgeon, perhaps as low as 2 to 3%. If bacterial endocarditis is already present when the diagnosis is first made, immediate treatment with penicillin should be instituted and operation carried out when the patient has responded and is in better general condition. If the response is not good, operation should not be deferred, as it was curative in some of these cases even before the introduction of penicillin, but the operative risk is then greater.

The routine operation should be ligation of the ductus, and the risk of recanalization is slight. Before carrying out the operation it is essential to make sure that the ductus is the only lesion. The slightest degree of cyanosis or of right ventricular preponderance in the electrocardiogram suggests some complication as well as a patent ductus, and the patient might then be made worse by its closure. Aortic regurgitation may sometimes simulate the condition.

Coarctation of the Aorta

This condition is still missed, as was shown by the finding of it in many men during or even at the end of their military service during the recent war. The auscultatory signs are often insignificant, though there are generally systolic murmurs which may be widespread and heard in the back over dilated collateral vessels.

The essential feature is the high pressure in the upper part of the body, with a lower pressure in the legs; and if the femoral pulse is felt for in all patients with high brachial blood pressure or with abnormal pulsation in the neck the diagnosis should not be missed. Dilated collateral vessels will then probably be found over the chest and back, especially round the scapula. These are often made more obvious if the patient bends forward with his arms hanging by his side. Notching of the ribs on the x-ray films may be further confirmation, but the diagnosis should be made first without this.

This condition can now be cured by surgery, but it is more difficult to decide whether operation is indicated, as the prognosis is often good and the operative risk is much greater. The rise of blood pressure is not progressive through life, though it may progress in childhood and may not reach its maximum figure till 12 or even 20 years of age. The patient with a blood pressure of 200 may show no more than a little left ventricular enlargement and may not become more handicapped in the course of another twenty years. Roughly speaking, the prognosis is good in these patients without symptoms in whom the lesion is found by routine examination after the age of 20, and not so good in those in whom symptoms develop in childhood, though very slight limitation on exertion or slight aching of the legs need not count in this direction.

Operation should be performed only by those with special experience, and even in such hands the mortality may be about 15%, though possibly the next few years may show some reduction in this.

If the patient has serious symptoms in childhood, or if the blood pressure continues to rise greatly after 14 or 15 years of age, operation should probably be advised. After 20 the risks of operation are much greater and an anastomosis of an aorta that already shows atheromatous change may be difficult or impossible, so that a greater degree of disability would be needed to justify the risks.

Aortic incompetence is a fairly common association and may be due to dilatation of the aorta or to changes in the aortic valves and may be responsible for some of the disability. Congenital cerebral aneurysms are often present in these cases and may lead to fatal subarachnoid haemorrhage, and though the risk of this is minimized by reduction of the blood pressure it is not entirely removed.

Pulmonary Valvular Stenosis

This may occur with Fallot's tetralogy but more often with a closed ventricular septum. The stenosis may not be severe in spite of striking physical signs—a rough systolic murmur and thrill, and a diminished second sound in the pulmonary area—and the patient may go through life somewhat but not greatly limited, and may remain acyanotic till the end. When the stenosis is more severe the course will depend on whether the foramen ovale is sealed when the pulmonary stenosis forces the pressure on the right side above that on the left. If it is sealed so that

there is no chance of a right-to-left shunt developing, the patient will remain acyanotic but the heart will hypertrophy greatly in an attempt of the right ventricle to overcome the stenosis.

If a patient in the "teens" or early twenties who has been acyanotic becomes cyanotic and much more disabled, the most likely diagnosis is pulmonary valvular stenosis with a right-to-left shunt through a patent foramen ovale.

Borderline Cyanosis

The diagnosis is difficult in this intermediate group, in which it is hard to decide if the patient is cyanotic or acyanotic, as so many possibilities have to be considered. The cause of the cyanosis may be peripheral. Some patients will become cyanotic in the course of a few years—for example, those with pulmonary valvular stenosis in whom the pressure on the right side is rising and forcing the shunt through a foramen ovale that was not sealed. Others will remain much the same without any increase of cyanosis until terminal heart failure, and these often have large septal defects. The only cases of immediate practical importance are those rare cases of Fallot's tetralogy in which the disability is great and the cyanosis trivial, probably because the pulmonary stenosis is severe and the aorta only slightly overriding: even then it is rare for the cyanosis not to be constantly present at rest.

Illustrated by Miss Sylvia Treadgold. This article will be concluded next week.

THE LEBANON HOSPITAL

The fifty-first annual meeting of the Lebanon Hospital for Nervous Diseases was held at Manson House, London, on April 28. This hospital was founded in 1900 at Asfuriyeh, on the slopes of Mount Lebanon, about six miles from Beirut. It was founded on the initiative of a Swiss missionary, and the money was raised in Europe, with a central committee in London. Britain has played a leading part in raising funds for the hospital and providing staff. Dr. William Ford Robertson, the new medical director, formerly director of the West of Scotland Neuropsychiatric Research Institute, recently took up his duties.

At the meeting the value of the work of the hospital was stressed by Sir Ronald Storrs, who presided, and by the Lebanese Minister in London, Dr. Victor Khouri. Dr. Khouri mentioned that the Lebanon itself was now contributing to the upkeep of the hospital, and a gift had been received from the Lebanese Government. Mr. Raymond Baldwin announced that it was intended to improve the reception block, build married quarters for the medical staff, and sink a new well.

Professor Alexander Kennedy presented the medical report for the past year, prepared by the senior resident physician, Dr. Manugian. Drawing attention to the variety of nationalities and religions among the 253 male and 187 female in-patients admitted during 1949, Professor Kennedy said that he doubted whether any hospital could be as free from prejudice of any kind. The Lebanon Hospital was regarded as the best of its kind in the Middle East. All modern treatments were used, and the results compared favourably with those of mental hospitals in Britain. He thought that the hospital would have a very important function in the training of doctors. It had been training nurses for some time, and he was disappointed that there had not been a better response from other mental hospitals in the Middle East to a circular calling attention to the existence of the nurses' training school. In addition to its in-patient facilities, the hospital ran a neuropsychiatric clinic in Beirut, in which there had been 2,121 consultations during the past year.